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Central Neurocytoma with Extensive Intratumoral Hemorrhage: A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 39-year-old
Final Diagnosis: Central neurocytoma (WHO Grade 2) with extensive intratumoral hemorrhage
Symptoms: Dizziness • headache • loss of appetite • presyncope
Clinical Procedure: Radiographic imaging • rehabilitation • tumor extirpation
Specialty: Neurosurgery • Radiology

Objective: Rare disease

Background: Central neurocytoma (CN) is a rare neuronal tumor of neuroepithelial origin. It has been assigned to World Health Organization classification grade 2. These tumors are usually benign and located in the anterior half of the lateral ventricle, though they can also be found in the third and fourth ventricles. Left untreated, a CN can cause blockage of cerebrospinal fluid, thus leading to hydrocephalus. CNs are exceedingly uncommon, making up just 0.1-0.5% of primary intracranial tumors. The tumors typically develop in people aged 20 to 40. There are no official guidelines on how to treat CN, so treatment options are often individualized on the basis of specific case findings.

Case Report: A 39-year-old man with an uncomplicated medical history presented with dizziness, increasingly worse headaches, presyncope, and a loss of appetite. Radiological data and postoperative histopathological and histochemical analysis led to the diagnosis of CN with extensive intratumoral hemorrhage. Surgical resection of the tumor was proposed to the patient, to which he agreed.

Conclusions: CN is a benign tumor, but it can cause serious or life-threatening complications. Gross total resection of the tumor is recommended if possible, and if deemed beneficial to the patient's clinical condition. This case reports the symptoms of a patient with CN, who underwent gross total resection and showed no sign of any residual tumor tissue on postoperative MRI. By reporting these types of cases, we can take necessary steps ahead of widespread agreement on optimal treatment of patients with neurocytomas.

Keywords: Neurocytoma • Neurosurgery • Radiology

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Background

Central neurocytoma (CN) is a rare neuronal tumor of neuroepithelial origin. It has been assigned to World Health Organization (WHO) classification grade 2 [1]. These tumors are usually benign and located in the anterior half of the lateral ventricle, though they can also be found in the third and fourth ventricles. Left untreated, such tumors can cause hydrocephalus, which is a blockage of the cerebrospinal fluid system [2]. CNs represent just 0.1-0.5% of all primary intracranial brain tumors [3,4]. The tumors typically develop in people aged 20 to 40. This applies to two-thirds of CN diagnoses, and there are no reported gender predilections in this regard [5]. Magnetic resonance imaging (MRI) can be the first step in a diagnosis, but only histopathological analysis can yield a definitive one [6]. There are no official guidelines on how to treat CN, and so treatment options are often individualized on the basis of specific case findings. Surgery is currently the cornerstone of CN treatment. This can involve gross total resection or subtotal resection. The prognosis for individuals who have undergone insufficient tumor resection can be improved with postoperative and adjuvant radiotherapy [7].

In the present paper, we present a case that illustrates and demonstrates key findings and treatment highlights related to a patient who was diagnosed with a CN with extensive intratumoral hemorrhage.

Case Report

A 39-year-old man visited the hospital, reporting dizziness, worsening headaches, presyncope, and a loss of appetite which had lasted for the past 4 days. The patient had no other past illnesses or family history of same. An initial workup involved a routine blood/biochemistry panel, neurological examination, and a non-contrast enhanced computed tomography (CT) scan of the patient's head. Neurological examination included assessment of the patient's mental status, motor function, and sensory function, and evaluation of reflexes. No alterations were found in the neurological assessment. There were also no major alterations in the blood workup. The CT scan revealed a mass with partly hemorrhagic formation that had a maximum diameter of 43 mm. It was found in the left lateral ventricle along with generalized cerebral edema with a midline shift to the right and a pronounced hydrocephalus in the lateral ventricles (**Figure 1**). The first CT scan characterized the finding as an intracranial mass with hemorrhagic component. The patient was hospitalized at the Department of Neurosurgery. Cranial MRI using a SIGNA™ Voyager 1.5T MRI machine was conducted the following day to reveal a large intraventricular neoplasm (CC 46×LL 35×AP 39 mm) with central hemorrhages in the vicinity of the foramen of Monro and in the left lateral ventricle. This was causing obstructive hydrocephalus and shifting the brain's midline structures (**Figures 2-4**). The contrast-enhancing component of the tumor on diffusion-weighted imaging (DWI) sequence showed restricted diffusion at an apparent diffusion coefficient (ADC) value of 1009×10^{-6} mm^2/s with a *b* value of 1000 s/mm^2. A differential diagnosis of choroid plexus papilloma and choroid plexus carcinoma were acknowledged in the MRI report's conclusion. Surgical resection was proposed to the patient for extirpation of the tumor (gross total resection) via osteoplastic trepanation. The patient agreed.

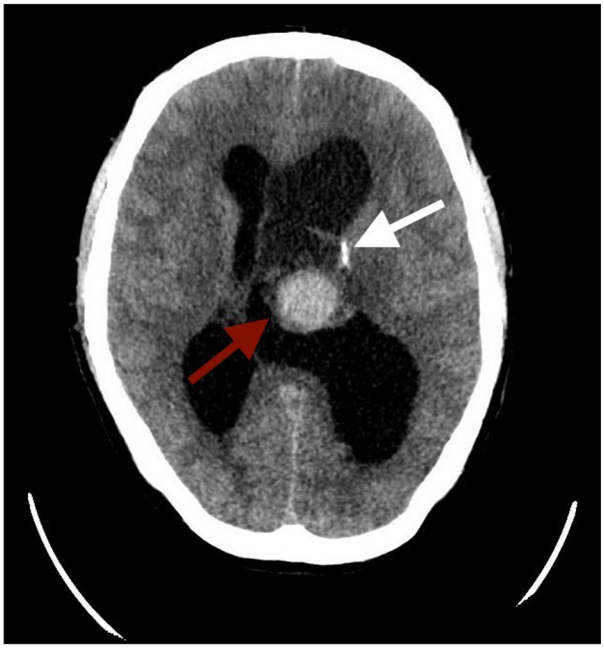

Figure 1. CT of the head, axial reconstruction, showing a mass of non-homogeneous density in the left lateral ventricle (red arrow), with a component of a hemorrhage. Focal calcification on the lateral side of the tumor can be seen (white arrow). CT – computed tomography.

Surgery and Pathology

The patient was anesthetized with general anesthesia including sevoflurane, and he was placed in the right lateral decubitus position. Osteoplastic trepanation was performed on the patient's left parietal bone. The dura mater was separated from the bone and cut open to expose the brain. A 2-cm cerebrotomy was performed in the middle level of the parietal lobe, thus entering the lateral ventricle, and, specifically, the junction of the parietal-occipital and the temporal horn. Abnormal tissue was found medially upon entering the lateral ventricle, and this was extirpated within the limits of visibility. The parietal horn of the lateral ventricle could be seen after the extirpation, and viscous cerebrospinal fluid was discharged. More tumor tissue was extirpated, and a local and

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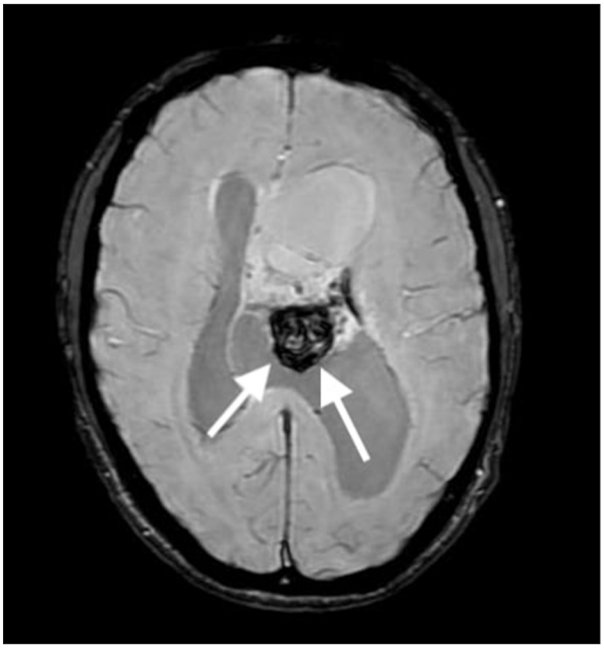

Figure 2. MRI of the head, SWI sequence, axial plane. A solitary lobulated mass is centered in the left lateral ventricle close to the foramen of Monro, with heterogeneous signal, visually well-demarcated, and with central hemorrhage (white arrows). MRI – magnetic resonance imaging; SWI – susceptibility weighted imaging.

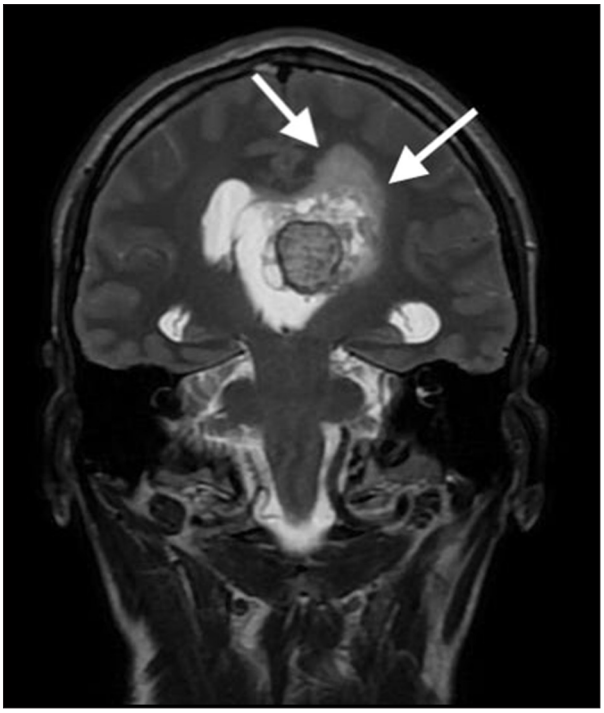

Figure 3. MRI of the head, T2WI sequence, coronal plane. There is edema in the periventricular white matter, around the left lateral ventricle (white arrows). MRI – magnetic resonance imaging.

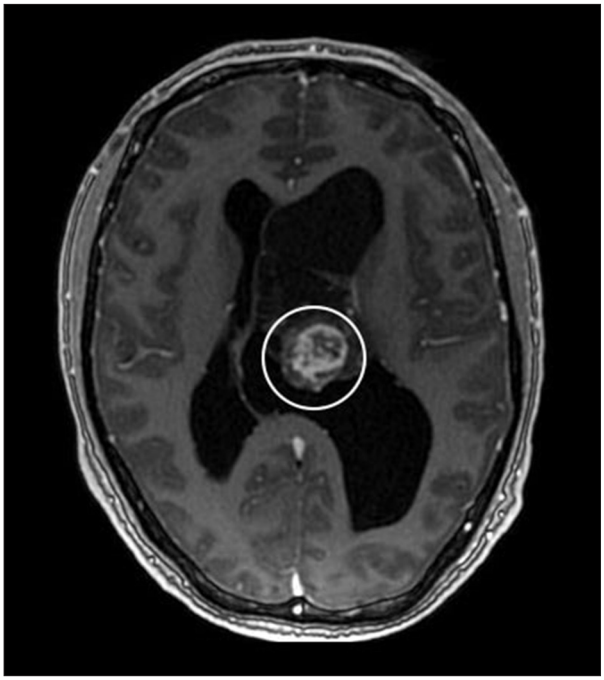

Figure 4. MRI post-contrast T1WI sequence of the head, axial plane. A central component of the tumor demonstrates intense enhancement in a point-like manner. MRI – magnetic resonance imaging.

topical hemostatic agent – surgical gel SNoW™ – was employed to achieve hemostasis. The dura mater was surgically closed, the bone cover was reinserted, and the subcutaneous and cutaneous layers of the skin were sutured. The patient was admitted to the Intensive Care Unit (ICU). The tumor tissue (2.4×1.7×1.6 cm) that was extracted during the surgery was sent for pathological analysis.

The histopathological analysis found a structure with very large areas of centralized hemorrhage in the tumor material. This formation had numerous blood vessels, massive hemorrhages, fibrin, and focal inflammatory cell infiltrate. A tiny amount of preserved tumor tissue was discovered around some of the micro-preparation edges throughout the hemorrhagic core portion of the formation. This tissue was composed of giant monomorphic cells with round, small monomorphic nuclei, and fine-grained chromatin. Along the nucleus we found many light "halo" zones. The tumor contained many tiny capillaries. There was no definitive evidence of mitotic activity, nor was there any obvious cell atypia. Histochemical analysis of the tumor tissue revealed a positive synaptophysin protein. Glial fibrillary acidic protein (GFAP) was not seen around the area of the tumor. IDH1 R132H enzyme, alpha-thalassemia/mental retardation syndrome, nondeletion type, X-linked (ATRX) and epithelial membrane antigen (EMA) were all negative. The Ki-67 proliferation fraction around the area of the tumor was low; only around 2%.

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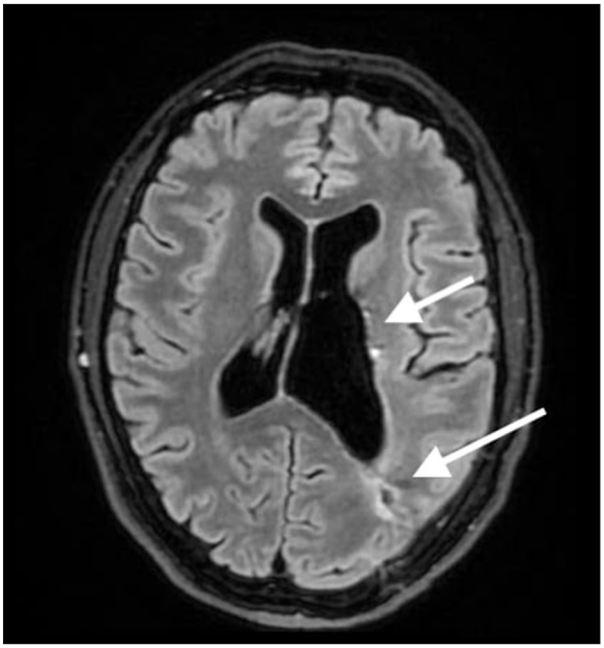

Figure 5. Postoperative MRI FLAIR sequence of the head, axial plane. No sign of any residual tumor tissue in the left ventricle or brain tissue can be seen. Postoperative scar tissue can be visualized (white arrows). MRI – magnetic resonance imaging; FLAIR – fluid-attenuated inversion recovery.

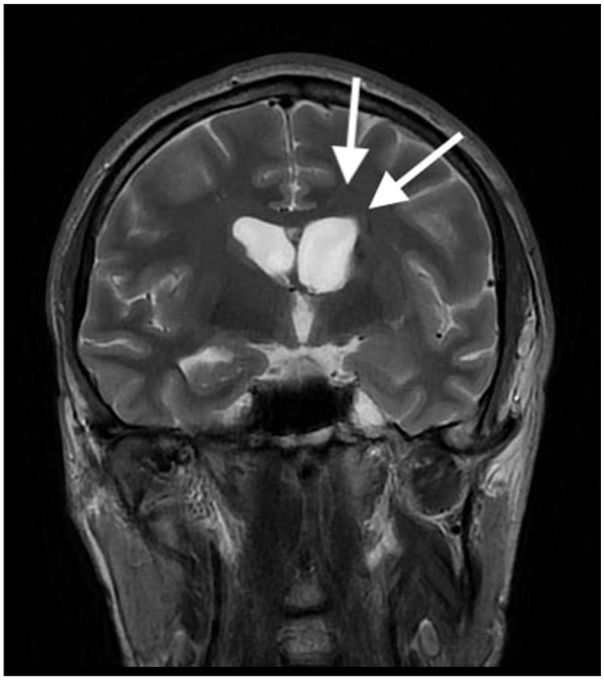

Figure 6. Postoperative MRI, coronal T2WI sequence of the head shows periventricular white matter edema that has considerably decreased (white arrows). MRI – magnetic resonance imaging.

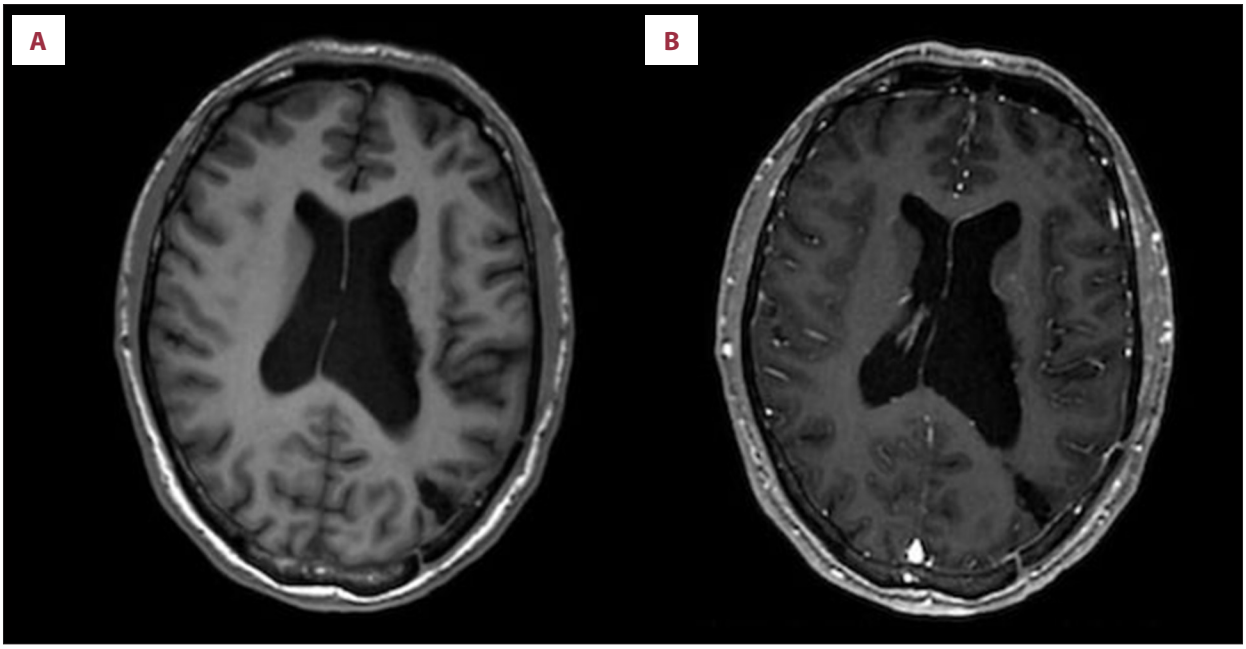

Figure 7. (**A**) Postoperative MRI of the head with pre-contrast T1WI sequence. (**B**) Postoperative MRI of the head with post-contrast T1WI sequence showed no abnormal enhancement, demonstrating no residual or recurrent tumor. MRI – magnetic resonance imaging.

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The diagnosis of CN (WHO grade 2) with extensive intratumoral hemorrhage was determined on the basis of morphological and immunohistochemical examination data, as well as radiological data that indicated a typical location for CN.

Outcome and Followup

The patient dynamically developed an acute onset of right-sided hemiparesis, as well as sensory and motor aphasia in the wake of the surgery. Both types of aphasia progressively improved during daily rehabilitation, which also improved the hemiparesis. The patient steadily regained strength and mobility on his right side, which had been affected. A CT scan performed after the operation showed no residual tumor mass, a shift of the remaining midline structures to the right, and residual hemorrhage in the right lateral ventricle. The patient spent 1 month at the rehabilitation department. Favorable dynamics were observed during the process. Independence of movement and daily tasks increased. A month later, the patient returned to the rehab department, complaining of memory loss, trouble walking, immobility in his right leg and arm, and difficulty in lifting objects weighing more than 1.5 kilograms. The patient underwent repeated rehabilitation with the goal of training for therapeutic exercises which he could perform himself. This included aerobics, strength training, and cognitive enhancement. After a followup visit 3 months after the surgery, MRI scans showed no residual tumor within the brain. Postoperative scar tissue and hemosiderin inclusions could be seen around the surgical site (**Figures 5-7**).

Discussion

CN was initially identified in 1982 by Hassoun et al [5]. Between 2004 and 2015, the National Cancer Database collected data from 223 404 brain tumor patients. Of these, 868 (0.4%) had been diagnosed with neurocytoma. The average age at the time of diagnosis was 31, and the average size of the tumor was 4-5 centimeters. There were 622 intraventricular tumors, 168 extraventricular tumors, and 78 cases in which the tumors were overlapping or not specified [8]. The age and tumor size of our patient appeared to correlate with these data. CN presentation with tumoral hemorrhaging is very rare, and very few cases have been documented [9-11].

Differential diagnosis of CN includes subependymomas, astrocytomas, meningiomas, choroid plexus papillomas, and malignant teratomas. Similar symptoms or patient outcomes to CN can result from these neoplasms [12,13].

In our case, both CT and MRI were put to use. Both medical imaging modalities helped to characterize the tumor and to identify its precise location. During a CT scan, CN usually appears as an iso- to hyper-dense or mixed-density mass which is frequently found inside the lateral ventricles and close to the septum pellucidum and the foramen of Monro [14]. Calcification of the tumor can also be seen on a CT scan. MRI imaging showed low T1 and high T2 signal intensity with calcification and variable contrast enhancement, with the tumors usually located at the lateral ventricle or without third-ventricle extension or attachment to the septum pellucidum [2]. The tumor showed restricted diffusion on DWI sequence, with an ADC value of 1009×10^{-6} mm^2/s and *b* value of 1000 s/mm^2. Susceptibility weighted imaging demonstrated intratumoral bleeding of the tumor. The main mass of the tumor demonstrated a cystic component, obstructing the foramen of Monro, enlarging the left lateral ventricle, and causing obstructive hydrocephalus, resulting in a shift of the midline structures of the brain.

There is no widespread agreement on how to treat neurocytomas. Surgical management with gross total resection is usually recommended as the primary therapy. This often involves positive prognoses and reduces the likelihood of recurring CN. If subtotal resection is the surgical method, then that decreases the rate of survival and increases the risk of relapse [15]. For patients who have undergone subtotal resection, postoperative adjuvant radiotherapy can improve the prognosis and the outcome [7]. In the present case, histopathological analysis did not indicate any sign of residual tumor tissue on the resection margin, which indicates that gross total resection was successful. The patient did not require any adjuvant type of radiotherapy. Some risks of toxicity have been associated with this type of therapy. The data do not show any statistically significant improvement in the rate of survival among patients who have undergone gross total resection or radiotherapy [16]. The overall risks outweighed the pros of adjuvant radiotherapy for our patient.

Positive therapeutic and prognostic characteristics of CN are significant and important talking points. Prognoses for CN are typically favorable, but of essential importance is a precise diagnosis, leading to prompt treatment.

Conclusions

We have presented a case of a rare CN tumor with extensive intratumoral hemorrhage. CN is a benign tumor which can cause serious or life-threatening complications. Gross total resection of the tumor is recommended if this is beneficial to the clinical condition of the patient. We have reported the symptoms of a patient with CN, who underwent gross total resection and showed no sign of any residual tumor tissue on postoperative MRI. By reporting these types of cases, we can take necessary steps forward towards developing widespread agreement on treatment of patients with neurocytomas.

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Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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